

(PANK2).² Interestingly, our patient had significant cognitive impairment and palatal tremor in addition to the movement disorders so far described in patients with neuroferritinopathy. In other neurodegenerative disorders, particularly HD, the causative proteins may be involved in iron metabolism.³ Thus, cognitive impairment may be predicted to occur in neuroferritinopathy, especially in the presence of a pre-existing hyperkinetic movement disorder.⁴

The development of palatal tremor in our patient deserves further explanation. Palatal tremor (previously known as palatal myoclonus) may be classified as essential or symptomatic.⁵ It is thought that palatal tremor arises because of functional disruption in "Mollaret's triangle", which consists of the inferior olivary nucleus, red and dentate nuclei. The symptomatic form is usually associated with hypertrophy of the inferior olivary nucleus and may arise from vascular lesions, particularly in the cerebellum.⁶ Further evidence for this hypothesis comes from a positron emission tomography study, which showed hypermetabolism in the inferior olivary nucleus.⁷ Most patients also have cerebellar ataxia. However, palatal tremor may also occur in other conditions including multiple system atrophy, progressive supranuclear palsy, and Alexander's disease.⁵ As in our case, symptomatic palatal tremor is not usually associated with ear clicking. Presumably, in our patient, iron deposition in the dentate nuclei was responsible for disruption of rubral and olivary pathways.

Ferritin is an iron storage protein and alteration in structure of the carboxy terminus could lead to the release of free iron and excessive oxidative stress.⁸ In other conditions, such as haemosiderosis, the use of iron chelators has been advocated as a potentially useful treatment. Results, in the main, have been disappointing. Whether free radical scavengers, such as idebenone,⁹ have useful therapeutic value in neuroferritinopathy remains to be seen.

Neuroferritinopathy should be considered in all patients with a hyperkinetic movement disorder, imaging evidence of iron deposition within the brain, and an autosomal dominant family history.

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Cocaine induced hypokalaemic periodic paralysis

The use of cocaine has been associated with a number of psychiatric, medical, and neurological complications. This is the second reported case of a patient who suffered three distinct episodes of paralysis after engaging in a cocaine binge.

Case report

A 33 year old male horse breeder with no significant medical history was evaluated at the Texas Tech Health Sciences Center after the abrupt onset of ascending generalised weakness. He reported not being able to walk or lift his arms or legs, much less climb up or get down the stairs of his home. He reported no bowel or bladder incontinence, loss of sensation, headache, nausea, or vomiting. The patient did report mild chest pain at the time. Ten days before his initial evaluation he had suffered a very similar episode but had not sought medical attention. At the time of his evaluation the patient stated that he would be better in 24-48 hours. A very similar event had occurred five years earlier, for which he was seen in an urgent care facility and discharged home; symptoms resolved after 2-3 days. Records of this first episode were not available, although he reported that potassium supplements were provided at that time.

Physical examination found an uncomfortable appearing, slightly dishevelled, unshaven man with no spontaneous motor activity. Vitals signs were a pulse of 88 beats/min, respiration 16 breaths/min, and blood pressure 132/94 mm Hg. Neurological evaluation found an awake, alert, and oriented person. Speech and language were normal. Cranial nerves were intact. Motor examination found normal bulk with a reduction in tone. Strength was 2/5 in all major muscle groups with a very mild left upper limb predominance. Neck extensors and flexors were 5/5. Bulbar muscles were spared. No myoedema, myotonia, fasciculations, or other abnormalities were noted. The sensory examination was normal and reflexes were symmetric with no Babinski signs. A complete blood count and comprehensive metabolic panel, including thyroid studies, urine drug screen, blood alcohol concentration, and erythrocytation rate, were performed. Cardiac enzymes were normal. Neuroimaging of the brain and spinal cord were normal. Forced vital capacity and negative inspiratory fraction were normal. Laboratory investigations showed a blood glucose concentration of 6.6 mmol/L, sodium 141 mmol/L, calcium 2.27 mmol/L, and creatine kinase (CK) 395 IU/L. Acetylcholine receptor antibodies were drawn at the time of

admission and subsequently shown to be in the normal range. Two laboratory investigations were of particular interest. The patient's potassium concentration was 1.9 mmol/L and urine toxicology screen found the presence of cocaine, cannabinoids, and benzodiazepines.

The patient had initially denied any illicit drug use but later admitted to having engaged in a cocaine binge the previous night and before the previous two episodes of weakness. There was no family history of periodic paralysis or other neuromuscular disorders. Supplemental potassium was provided and the patient's strength gradually improved with rising concentrations of serum potassium. Nerve conduction studies and electromyography were normal at 48 hours after the onset of symptoms. He was discharged home to an outpatient substance abuse program three days later with almost complete resolution of symptoms. At the time of discharge, the serum potassium concentration was 4.5 mmol/L and the CK concentration declined to 133 IU/L.

It is not clear why the use of cocaine led to such severe generalised weakness and hypokalaemia in this patient. Nalluri *et al*¹ reported a similar case and suggested that the hypokalaemia was caused by an intracellular shift of potassium secondary to the adrenergic effects of cocaine; a hyperadrenergic cause of periodic paralysis in patients suffering from thyrotoxicosis has also been postulated. In their report, as in this case, the patient responded quickly to potassium supplementation. An alternative mechanism may have been cocaine's potential effects on potassium channels.^{2,3} The increased CK and serum glucose concentrations were felt to be the result of cocaine's effects.

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Competing interests: none declared.

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Sulcal abnormalities on brain magnetic resonance imaging in the Guillain-Barré syndrome

The Guillain-Barré syndrome is an immunologically mediated condition affecting the peripheral nervous system. There is evidence that Guillain-Barré syndrome, Miller-Fisher syndrome, and Bickerstaff brain stem encephalitis form a closely related spectrum of disorders.¹ Magnetic resonance imaging (MRI) abnormalities in the spinal cord in these conditions have been well described,² but intracranial findings are infrequent. We report resolution of sulcal changes on serial MRI of the brain concomitant with clinical recovery in a typical case of Guillain-Barré syndrome.

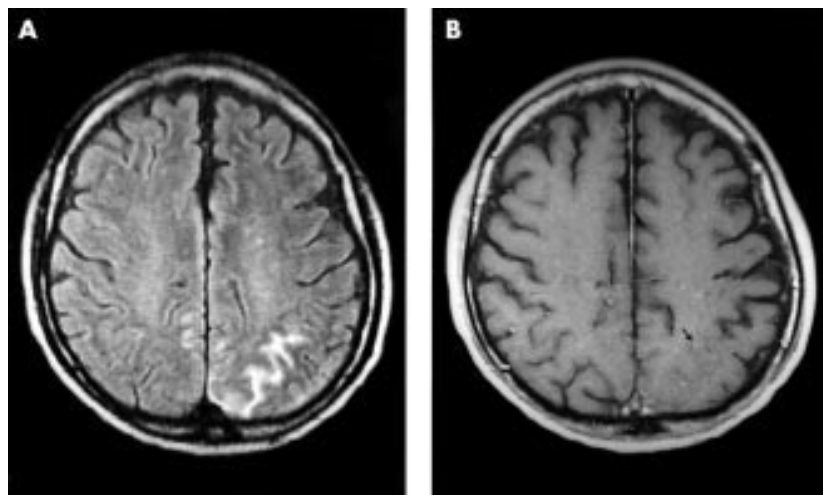


Figure 1 (A) Axial FLAIR (TR 9000/TE 110/TI 2500 ms) section through the centrum semiovale above the level of the lateral ventricles shows abnormal high signal within the left parietal sulci, without intracerebral vasogenic oedema. (B) Corresponding axial enhanced T1 weighted section (TR 540/TE 12 ms) showing obliteration of the normally dark CSF containing sulci (compare with frontal sulci) and subtle enhancement (indicated by arrows).

An 81 year old man had a one week history of progressive lower limb weakness and numbness associated with pain radiating down his right leg. There was no preceding history of infection or trauma. He had no significant past medical history.

On examination, he was alert and rational. There was no slurring of speech or paresis of the extraocular muscles. Cranial nerve and visual field examination was unremarkable and the neck was supple. He had mild proximal symmetrical upper limb weakness (MRC grade 4/5) and bilateral lower limb weakness (MRC grade 3/5). He had difficulty in walking unaided and in tandem walking. Sensory loss to touch was elicited in the distal lower extremities in stocking distribution. Reflexes in all four limbs were absent. The clinical features were consistent with the Guillain-Barré syndrome.

The patient was initially referred to an orthopaedic surgeon for possible lumbosacral spondylitic disease. Unenhanced MRI scans of the lumbar and thoracic spine showed mild degenerative changes and excluded intrinsic cord abnormalities or external compression. Subsequent enhanced cervical spine MRI scans were also negative. He was then referred for a neurological opinion.

Nerve conduction studies revealed significantly prolonged distal motor latency (median motor distal latency 6.7 to 9.8 ms; posterior tibial distal latency 7.7 to 9.4 ms) and reduced conduction velocities in the median (40.3 m/s), ulnar (39.6 m/s), and tibial nerves (35.7 m/s). F responses were prolonged (> 37 ms) or absent in all four limbs. Cerebrospinal fluid examination showed absent cells with raised protein of 0.8 g/L, normal glucose concentration, and positive globulin. Bacterial culture and viral studies were negative.

The first MRI of the brain, obtained to exclude a central cause for weakness and gait abnormalities during the first week of admission, showed left parietal and superior occipital sulcal hyperintensities on the fluid attenuated inversion recovery (FLAIR) sequences, in addition to subtle enhancement with contrast administration (fig 1). A repeat MRI one week later showed mild improvement. A third MRI two months after initial presentation showed resolution of the focal abnormalities.

In the related Miller-Fisher syndrome, MRI changes have been documented in the cranial nerves,³ spinocerebellar tracts, and pons.⁴ We postulate that our MRI findings represent a focal manifestation of a wider immunologically mediated reaction within the subarachnoid cerebrospinal fluid bathed space. This focal sulcal reaction probably represents a local concentration of proteinaceous fluid and correlates well with the CSF findings of high protein levels but an absence of cells.⁴ These MRI changes resolved with immunoglobulin treatment and clinical recovery. While MRI changes have been reported in the subcortical regions in demyelinating neuropathies, most probably from focal demyelination, sulcal changes have not been described. Serial MRI studies are a sensitive technique for documenting cerebral cortical abnormalities in this condition, even in the asymptomatic setting as demonstrated here.

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Muscle tissue oxygenation as a functional tool in the follow up of dermatomyositis

Near-infrared spectroscopy (NIRS) is a direct, non-invasive optical method for measuring local oxygenation and haemodynamics in muscle tissue. Although measurement of local oxygenation by NIRS has been used for the diagnosis of metabolic myopathies, the technique has not previously been applied to inflammatory myopathies. Dermatomyositis is a muscle disorder characterised by complement mediated capillary necrosis, resulting in ischaemia and hypoperfusion. We have now employed NIRS to study the effect of corticosteroid treatment on haemodynamics in muscle tissue in dermatomyositis.

The pathological features of dermatomyositis are characterised by a decreased number of capillaries per muscle fibre and necrosis of single muscle fibres or clusters of fibres at the periphery of the fasciculi.¹ Muscle fibre regeneration and an increased number of capillaries have been shown in dermatomyositis after intravenous immune globulin treatment,² but corticosteroids are still considered to be the first line of therapy. In the clinical setting, the effect of treatment is mainly assessed by muscle strength and creatine kinase (CK) levels. Direct measurement of capillary and muscle fibre status can only be done by repeated muscle biopsies. However, apart from the fact that muscle biopsies are invasive, they are also a static representation of muscle tissue at a fixed time point and at a particular location (selection bias).

This is the first time that NIRS, a non-invasive optical method for the measurement of oxygenation and haemodynamics in muscle tissue, has been used to study the effect of treatment in a patient fulfilling the clinical and histological criteria of definite dermatomyositis.³ A young woman from Aruba, aged 24 years, presented with subacute erythema of the facial skin and severe proximal muscle weakness (arm muscles: mean Medical Research Council (MRC) grade 3; leg muscles: MRC grade 2). Serum CK levels were slightly increased (220 IU/L). Five weeks after the onset of symptoms, treatment with corticosteroids was started in our department at a dose of 60 mg/day (for six weeks), the dose being subsequently tapered. CK levels decreased and muscle strength increased (arm muscles: MRC grade 4; leg muscles: MRC grade 3) in week 12.

Tissue oxygenation was measured by NIRS immediately before treatment was begun and again after three and seven weeks of treatment. NIRS is based on the relative tissue transparency to light in the near-infrared region, and on the oxygen dependent absorption changes of haemoglobin and myoglobin. Using a modification of the Lambert-Beer law, in which physical path length is incorporated to account for light scattering, it is possible to calculate quantitative values for oxygen consumption and blood flow in skeletal muscle. NIRS is non-invasive and measures oxygenation directly in the muscle. Moreover, it is relatively inexpensive, easy to apply, and applicable at the bedside.

In this study, NIRS measurements were obtained using a continuous wave near-infrared spectrophotometer (Oxymon, Biomedical Engineering Department, University